NHS England

Protocol for management of patients with pineal region tumours v1

West Midlands Cancer Alliance

Coversheet for Cancer Alliance Expert Advisory Group Agreed Documentation

This sheet is to accompany all documentation agreed by the West Midlands Cancer Alliance Expert Advisory Groups. This will assist the Clinical Network to endorse the documentation and request implementation.

EAG name	Brain and Central Nervous System	
Document Title	Protocol for management of patients with pineal region tumours	
Published date	April 2018	
Document Purpose	This guidance has been produced to support the management and follow-up of adult patients with pineal region tumours.	
Authors		
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Approval Signatures:	**EAG Chair**  Date: 20 April 2018	**Network Clinical Director**  Date: 20 April 2018

PROTOCOL FOR MANAGEMENT OF PATIENTS WITH PINEAL REGION TUMOURS

Date Approved	**April 2018**
Date for Review	**April 2021**

DOCUMENT HISTORY

Version	Date	Summary
1	April 2018	Reviewed by Cancer Alliance Expert Advisory Group

Scope of the Guideline

This guidance has been produced to support the management and follow-up of adult patients with pineal region tumours.

2. Background

2.1 Pineal region tumours are uncommon. The occur most frequently in paediatric patients and are rare in adult patients.

2.2 Several different tumour types can occur (Table 1)

2.3 Some tumour types respond extremely well to radiotherapy and/ or chemotherapy, and major surgical resection should be avoided in these patients.

2.4 Hydrocephalus frequently occurs and CSF diversion might be necessary even though the tumour bulk has been removed.

2.5 The British Neuro-oncology Society, in collaboration with the national Cancer Action Team, have produced guidelines for the management of pineal region tumours, and this pathway document is based on those guidelines.

Table 1: Pineal Region Tumours

Group	Subtypes
Germ cell tumours	Germinoma Non-germinomatous germ cell tumour Teratoma Embryonal carcinoma
Pineal tumours	Pineocytoma Intermediate differentiation pineal tumours Pineoblastoma
Astrocytic tumours	Tectal plate tumours High-grade gliomas Low-grade gliomas
Meningeal tumours	Meningioma
Others	Pineal cysts Dermoid cysts Lipomas Vascular lesions

3. Guidelines

3.1 Unless contraindicated, all patients should be investigated with MR scanning with view in 3 planes, and spinal imaging.

3.2 Blood and CSF tumour markers should be assayed. In most cases CSF can be obtained by lumbar puncture, but if contraindicated due to a mass lesion, obtaining ventricular CSF at the time of CSF diversion should be considered.

The relevant tumour markers are:

α-Fetoprotein (AFP)	- germ cell tumours
β-Human chorionic gonadotrophin (HCG)	- germ cell tumours
Carcino-embryonic antigen (CEA)	- teratomas
Placental alkaline phosphatase (PAL)	- germinomas.

3.3 Patients should be discussed in the Brain and Other Rare CNS Tumour MDT meeting after initial investigation (radiology & biochemistry), before surgery and after histological diagnosis. Discussion with a paediatric MDT dealing with pineal tumours should be considered.

3.4 A third ventriculostomy is the procedure of choice if CSF diversion is necessary.

3.5 A histological diagnosis is not necessary if a definitive diagnosis of a germ cell tumour can be made on tumour marker assay.

3.6 For other patients, resection should be performed by surgeons experienced in pineal region surgery. Needle biopsy should be avoided because of the proximity of large veins.

3.7 Patients with germinomas and pineoblastomas should be referred to a centre regularly performing craniospinal irradiation.

3.8 The standard radiotherapy dose for germinoma is craniospinal radiotherapy 24Gy in 15 fractions with a primary tumour boost of 16Gy in 10 fractions.

3.9 Patients with pineoblastomas should be managed according to the primitive neuro-ectodermal tumours (medulloblastoma) pathway

3.10 Patients with secreting germ cell tumours should be considered for chemotherapy with a combination of Cisplatin, Etoposide and Ifosfamide (PEI).

3.11 Patients found to have incidental pineal region cysts, without CSF pathway obstruction, and where the neuroradiologists are confident there are no features suggesting malignancy nan be managed by serial imaging with scans at 6 months, 12 months and 2 years.

4. Flowcharts